International Journal of Surgery 7 (2009) 559–560

Contents lists available at ScienceDirect

International Journal of Surgery

journal homepage: www.theijs.com

Adult presentation of giant retroperitoneal cystic lymphangioma: Case report

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ARTICLE INFO

Article history:
Received 2 September 2009
Accepted 16 September 2009
Available online 6 October 2009

ABSTRACT

This case describes the rare presentation of a retroperitoneal cystic lymphangioma in a 35 year old female patient. The lymphangioma ultimately progressed to the point of inducing clinical symptoms, thus requiring surgical removal – which was accomplished without incident. The relevant clinical pictures are included for educational value.

1. Case report

A 35 year old female presented with a 2–3 month history of right upper quadrant pain, early satiety, abdominal fullness and change in bowel habits. Suspecting the possibility of gallbladder disease, an abdominal ultrasound revealed was obtained. This revealed a complex, cystic mass of unclear origin abutting the liver. An abdominopelvic CT scan confirmed a large mass occupying the retropancreatic region and right retroperitoneum (Figs. 1 and 2). Cystic lymphangioma was felt to be the most likey diagnosis, and the patient was subsequently scheduled for exploratory laparotomy.

Upon entering the abdomen the cystic lesion was easily visible through the omentum and the retroperitoneal tissues. The mass was displacing the right colon medially and inferiorly, and the duodenum and pancreas anteriorly. After mobilizing the hepatic flexure inferiorly, a generous Kocher maneuver was used to expose the mass (Fig. 3). The mass was carefully dissected from the inferior vena cava, the right kidney, the right gonadal vein, and the duodenum. Due to extension of the mass under the portal structures, an open cholecystectomy was undertaken to allow delivery of the mass under the portal vein. The gastrohepatic ligament was then opened and dissection of the lymphangioma was continued down to just lateral of the celiac trunk, where it terminated. A harmonic scalpel was then used to transect the mass at its origin, allowing its complete removal (Fig. 4). Pathologic examination confirmed the diagnosis of a benign cystic lymphangioma (Fig. 5). Her postoperative course was uneventful and she was discharged home on post-op day 6.

2. Discussion

Cystic lymphangiomas are extremely rare, benign tumors of the lymphatic system. They are more common in the pediatric population, making this case somewhat unique. Additionally, only 1% of all lymphangiomas are reported to be retroperitoneal in location. Intra-abdominal lymphangiomas, which may arise from the mesentery, greater omentum, or retroperitoneum, are usually asymptomatic and found incidentally on imaging studies.[1] They may become symptomatic if they become large enough to impose on surrounding structures, as was the case in this patient.

Fig. 1. Coronal section of abdominal CT demonstrating a large water density occupying the retropancreatic region and much of the right retroperitoneal space.

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doi:10.1016/j.ijsu.2009.09.005

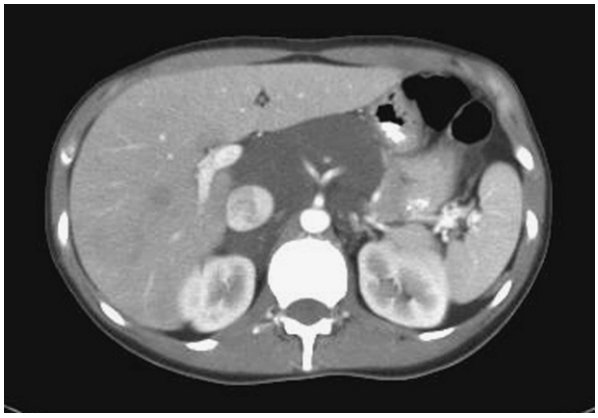

Fig. 2. Abdominal CT showing cystic massin the retroperitoneal space. Note displacement of retroperitoneal structures.

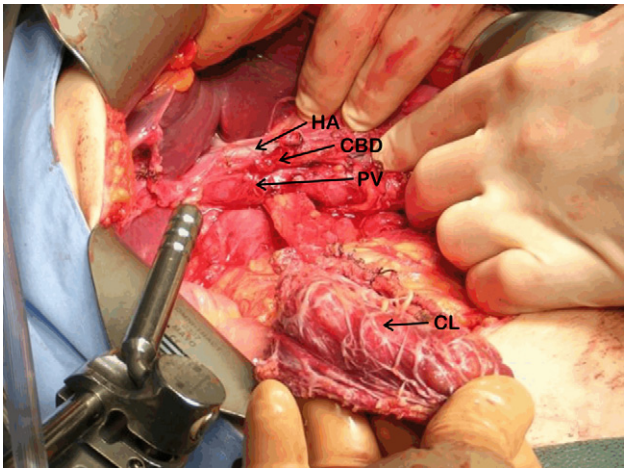

Fig. 3. Intra-operative photo demonstrating relationship of mass to biliary hilar structures. (**CBD** – common bile duct, **PV** – portal vein, **HA** – hepatic artery, **CL** – cystic lymphangioma).

Surgery is often required to ameliorate symptoms and for definitive diagnosis, since imaging studies are often unable to differentiate lymphangiomas from other cystic growths.[2] Complete excision is recommended to prevent progressive growth, infection, bleeding, and rupture.[3]

Conflict of interest
None declared.

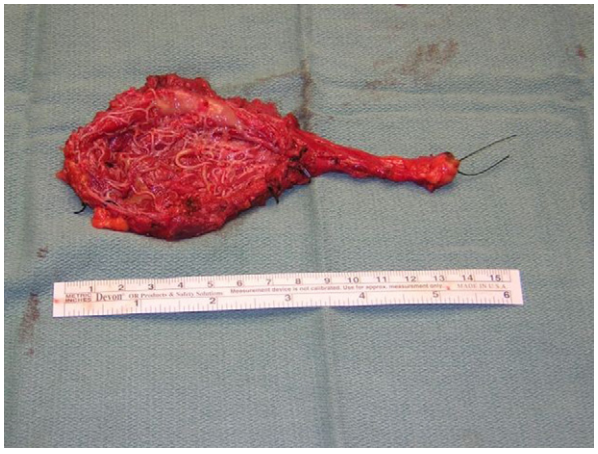

Fig. 4. Gross photograph of resected specimen.

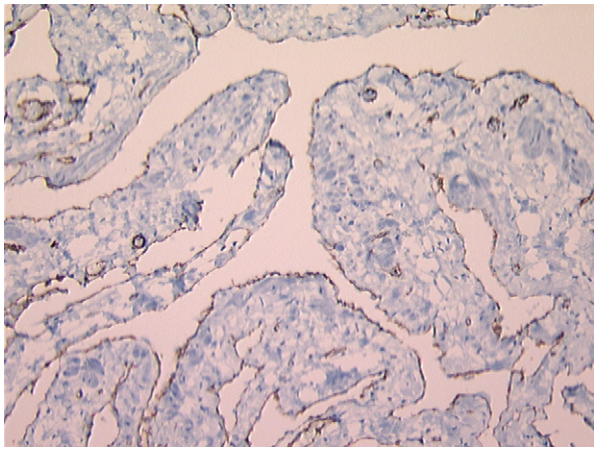

Fig. 5. Photomicrograph of cystic lymphangioma. (CD31 immunostain showing positive staining of the endothelial cells.) (200X).

Funding
None.

Ethical approval
None.

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